

## T helper IL17

**Utility:** Diagnostic screen for hyper – immunoglobulin E syndrome (HIES).

**Specimen:** 4 – 10 mL peripheral blood in sodium heparin (green top).

**CPT codes:** 86359, 86360, 86355, 86357, 88184, 88185 x 6, 81887.

### Clinical indication/general description

HIES, also known as Job's syndrome, is characterized by pulmonary infections, staphylococcal abscesses, eczema and abnormalities of bone and connective tissue. IgE levels typically are very high. HIES syndrome can look very similar to severe eczema, thus a laboratory test to differentiate these syndromes is clinically useful. The defects in Hyper-IgE syndrome are caused by mutations in the transcription factor *STAT3*. *STAT3* is required to induce CD4+ T cells to produce IL-17, a cytokine that is important for the elicitation of an effective immune response to several bacteria and fungi. The T helper IL-17 functional assay measures the ability of CD4+ T cells to make IL-17, which is defective in patients with HIES.

### Detection methodology

Peripheral blood mononuclear cells (PBMCs) are activated *in vitro* with PMA and ionomycin to induce the expression of IL-17 in normal T helper cells (CD3+CD4+), which is measured by flow cytometry using antibody that specifically recognizes IL-17. Simultaneously, IFN- $\gamma$  is measured within the T cytotoxic cell (CD3+CD8+) as a control to ensure adequate activation of T cells. An extremely low percentage of IL-17+ CD4+ T cells is associated with HIES, whereas the percentage of IL-17+ CD4+ cells is normal or slightly reduced in eczema.

